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## Psammocarcinoma of the ovary: a case report and review of the literature

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**Abstract** Psammocarcinoma is a rare form of serous carcinoma of the ovary, associated with extensive psammoma body formation and invasion of surrounding structures. We report the case of a 63-year-old woman who presented with a highly calcified abdominopelvic mass and a raised CA-125 marker and who, following a full staging laparotomy, was diagnosed with stage IIIB ovarian psammocarcinoma. Serous ovarian psammocarcinoma is characterized by massive psammoma body formation, and despite the limited number of cases reported, it appears that the clinical prognosis is much more favorable than for the usual serous carcinomas and is similar to that of serous borderline lesions of the ovary. A summary of all the reported cases is provided to highlight the clinical and prognostic features of this rare tumor.

**Keywords** Psammocarcinoma · Psammoma bodies · Serous carcinoma · Ovarian cancer

### Introduction

Psammocarcinoma is a rare variant of serous carcinoma of the ovary, characterized by massive psammoma body formation and low-grade cytological features. For the diagnosis of psammocarcinoma to be made, it is suggested that histologically the tumor should show psammoma bodies associated with 75% or more of the epithelial cell clusters and no more than moderate cytological atypia, essentially having characteristics similar to those of a borderline serous tumor. There should be tissue and/or vessel invasion and no solid cell sheets. If these criteria are present, it is claimed that the prognosis of these tumors

appears to be very good, with progression in only a very small minority of cases. A further diagnostic criterion that has been added requires that the cell clusters be no more than 15 cells in diameter, although the evidence that this is important to the outcome is essentially lacking [1].

### Case report

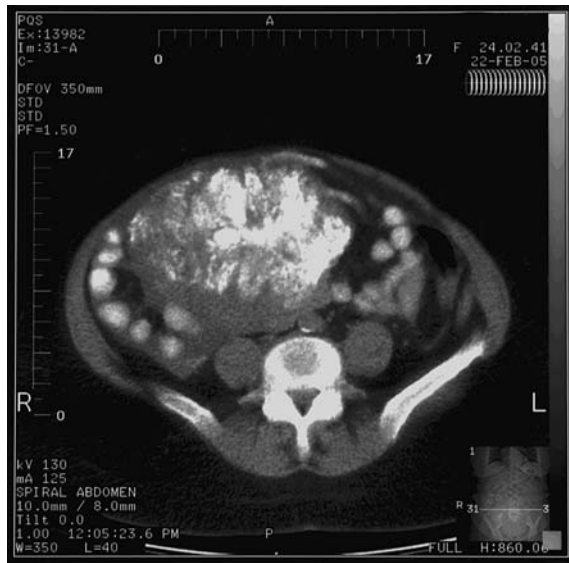
A 63-year-old woman was referred to our department with a large pelvic mass. She initially presented with a 3-month history of abdominal discomfort and increasing abdominal girth. Sonographic evaluation and computed tomography demonstrated a heavily calcified abdominopelvic mass that was initially suspected to be a leiomyoma (Fig. 1). A raised cancer antigen (CA)-125 level of 1,133 u/ml raised a strong possibility of an ovarian neoplasm.

The patient underwent a laparotomy. Intraoperative findings included 500 ml of ascitic fluid and a 10×15×9-cm right ovarian mass. The left ovary, both fallopian tubes, the uterus, and the omentum appeared normal. There were numerous tumor implants at the peritoneal surface but no evidence of upper abdominal disease or paraaortic or pelvic lymphadenopathy. A subtotal hysterectomy, bilateral salpingo-oophorectomy, and supracolic omentectomy were performed. The tumor was optimally debulked with no residual disease left at the end of the procedure apart from small seedlings in the pouch of Douglas.

On macroscopic examination, the neoplasm showed calcified solid areas and cystic spaces with small external and internal papillary excrescences. Multiple tumor nodules (<1.0 cm) involved the omentum. The left tube and ovary appeared normal, and the uterus measured 73×48×60 mm.

Histologic examination revealed a right ovarian tumor with numerous (>75%) psammoma bodies surrounded by a complex epithelial proliferation of papillae lined by a single layer of cytologically bland columnar epithelium (Fig. 2). Neoplastic elements included mild to moderate nuclear atypia with a high nuclear/cytoplasm ratio. No mitotic figures were identified. Occasional psammoma

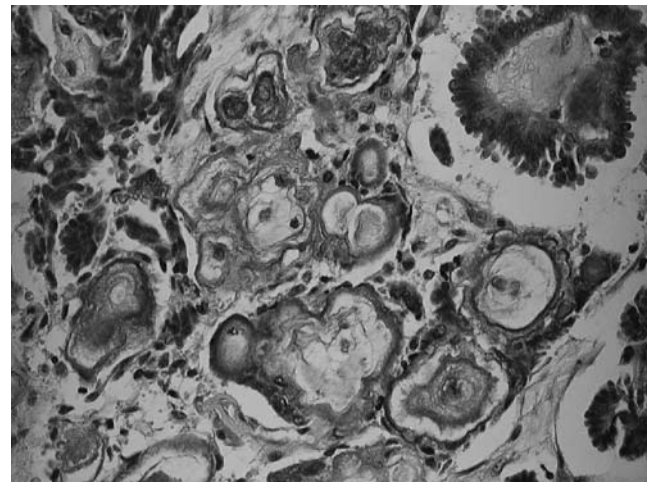
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**Fig. 1** Computed tomography scan showing a highly calcified pelvic mass

bodies were present in the parametrium with occasional epithelial clusters. The cytology of the peritoneal fluid was positive for adenocarcinoma cells. The endometrium showed a benign polyp, and foci of adenomyosis were seen in the myometrium. The endocervix was unremarkable.

The final pathological diagnosis was consistent with an International Federation of Gynecology and Obstetrics (FIGO) stage IIIB ovarian psammocarcinoma. The case was discussed at the multidisciplinary team meeting, and it was felt that adjuvant treatment with chemotherapy was indicated. The patient had six cycles of carboplatin.



**Fig. 2** Numerous psammoma bodies surrounded by groups of mildly atypical papillary epithelial cells (magnification  $\times 20$ )

## Discussion

Psammoma bodies are microscopic, laminated, calcified, extracellular bodies, thought to be the result of hydroxyapatite accumulations in cells that undergo degeneration. They are commonly found in several human tumors, including thyroid, meningeal, ovarian, and gastric neoplasms and duodenal carcinoids. In ovarian neoplasms, the presence of psammoma bodies is not limited to carcinomas but can also be found in benign and borderline serous epithelial tumors.

In 1990, Gilks et al. described eight peculiar primary ovarian neoplasms and three lesions of the omentum, which were called psammocarcinomas because of the massive psammoma body formation [1]. The other microscopic criteria proposed by Gilks et al. for diagnosing this tumor are the following: 1) destructive invasion of the

**Table 1** Reported cases of ovarian psammocarcinoma (LSO left salpingo-oophorectomy, LFU lost to follow-up, BSO bilateral salpingo-oophorectomy, NED no evidence of disease, TAH total abdominal hysterectomy, chemo chemotherapy, oment omentectomy, DFD death from disease)

Authors	Cases	Age (years)	Treatment	Stage (FIGO)	Follow-up
Gilks et al. [1]	1	66	LSO	IIIB	LFU
Gilks et al. [1]	1	72	BSO	IIIA	NED
Gilks et al. [1]	1	55	TAH, BSO, chemo	IIIA	NED
Gilks et al. [1]	1	36	TAH, BSO	IIIB	NED
Gilks et al. [1]	1	53	TAH, BSO, oment	IIIB	NED
Gilks et al. [1]	1	53	TAH, BSO, oment	IIIB	LFU
Gilks et al. [1]	1	76	BSO, oment	IIIB	NED
Gilks et al. [1]	1	59	LSO	IIIC	DFD
Kelley et al. [2]	1	18	TAH, BSO, oment, appendectomy, sigmoid colectomy, chemo	IIIC	NED
Pakos et al. [3]	1	49	BSO	IA	NED
Powell et al. [4]	1	59	TAH, BSO, oment, lymphadenectomy, appendectomy	IIIB	NED
Poggi et al. [5]	1	66	BSO, oment	IIIB	LFU
Cobellis et al. [6]	1	48	TAH, BSO, oment	IIIA	NED
Giordano et al. [7]	1	66	TAH, BSO, oment, chemo	IIIB	NED
Present case	1	66	TAH, BSO, oment, chemo	IIIB	NED

ovarian stroma, vascular invasion, or intraperitoneal viscera in extraovarian tumors, 2) no more than moderate cytological atypia, 3) no areas of solid epithelial proliferation except for occasional nests not more than 15 cells in diameter, and 4) at least 75% of papillae and nests associated or completely replaced by psammoma body formation. The prognostic importance of psammoma bodies in serous adenocarcinomas of the ovary had been unclear. However, some authors have noted that the presence of numerous psammoma bodies is associated with a better prognosis of the ovarian tumors.

Since then, five more ovarian cases have been reported [4–9]. The clinical and demographic data are listed in Table 1.

Patients ranged in age from 18 to 76 years. All but one patient [5] presented with stage III disease. Regarding prognosis, follow-up greater than 12 months was available in nine cases. Among these patients, eight were alive with no evidence of disease. The fact that only one of these patients (who had extensive, incompletely resected, intraperitoneal tumor with lymph node involvement) had died of their disease suggests a better prognosis for this type of tumor compared with the serous carcinomas of the usual type.

In summary, ovarian psammocarcinomas are rare tumors, and from the limited cases reported in the literature, their clinical prognosis appears to be much more favorable than that of the usual serous carcinomas and similar to that

of serous borderline lesions of the ovary. This tumor should be included in the differential diagnosis of pelvic masses, such as serous carcinoma and calcified leiomyoma.

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